The Fontan procedure in Australia: A population-based study

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Since its first description in 1971, the Fontan procedure has become the final stage of palliation for children not considered candidates for a two-ventricle repair.[1] The long-term future of these patients is uncertain, but it is clear that a large number of them will experience failure of their "Fontan circulation." The proportion of these patients who will eventually require heart transplantation is still unknown and may be altered by the use of emerging medical therapies and by surgical options such as the Fontan conversion.[2] The burden of this new patient population on health systems is difficult to evaluate, but it is reasonable to expect that most of them will require expensive therapies after their third decade with a Fontan circulation. Because the Fontan procedure is offered to such a heterogenous group of patients, it is difficult to evaluate the size of this patient population. The perioperative mortality of this procedure is now minimal, and the number of patients operated on will be the main determinant of the size of the population surviving with a Fontan circulation. In the past 3 decades, the Royal Children's Hospital has performed all Fontan operations in children from four Australian states; this has allowed us to evaluate the incidence of the Fontan procedure in our Australian population.

Patients and Methods

The first Fontan procedure performed in the Royal Children's Hospital in Melbourne was done in 1980. From that date to now, all patients born in Victoria, South Australia, Northern Territory, or Tasmania having a congenital heart defect and requiring complex surgery were referred to us. We reviewed demographic data from our surgical database to identify the state of origin of patients undergoing the Fontan operation. We estimated that most of the patients operated on in that time period would be born between 1970 and 1999. We then collected from the Australian Bureau of Statistics the number of live births in the four states between 1970 and 1999. Patients from each state were grouped by the year of their birth. The incidence of Fontan procedures per number of live births was calculated for each state by 5-year periods. Incidences were compared by the Fisher exact test.

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Received for publication June 7, 2007; accepted for publication June 21, 2007.

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J Thorac Cardiovasc Surg 2007;134:1353-4

0022-5223/$32.00

doi:10.1016/j.jtcvs.2007.06.035

Results

Between 1980 and 2006, a total of 356 patients underwent a Fontan procedure in the Royal Children's Hospital. Two hundred forty-seven of the patients who were born between 1970 and 1999 in Victoria, South Australia, Northern Territory, or Tasmania constitute the core of this study. Patient characteristics are given in Table 1. The overall incidence of patients ultimately requiring a Fontan procedure was 8.9 per 100,000 live births for the entire study period for the four states. Data by state for each 5-year period are given in Table 2; the incidence of the Fontan procedure in the four states did not differ significantly from the overall cohort. There were fewer Fontan procedures performed in the first decade than in the rest of the study period (5.7 vs 12.1 per 100,000; $P < .001$).

Discussion

The number of Fontan procedures performed per number of live births is now approximately 10 per 100,000 in our region. This number is unlikely to decrease greatly in the future. More parents may opt to interrupt pregnancies of fetuses diagnosed with heart defects; however, better management of babies born with major lesions, such as hypoplastic heart syndrome, has increased the number of potential candidates for Fontan surgery.

TABLE 1. Characteristics of 247 Fontan patients born in 1970 to 1999

Age at Fontan in years, median (IQR)	5.0 (3.2, 7.0)
Morphology, No. (%)	
Tricuspid atresia	55 (22)
Double-inlet left ventricle	62 (25)
Double-outlet right ventricle	43 (17)
Complete atrioventricular canal	22 (9)
Pulmonary atresia with intact ventricular septum	18 (7)
Hypoplastic left heart syndrome	15 (6)
Straddling atrioventricular valve	10 (4)
Other	22 (9)
Sex, No. (%)	
Male	143 (58)
Female	104 (42)
State, No. (%)	
Victoria	166 (67)
South Australia	63 (26)
Tasmania	12 (5)
Northern Territory	6 (2)

IQR, Interquartile range.

TABLE 2. Incidence of the Fontan procedure: Number of operations/number of live births (rate per 100,000 live births), 1970 to 1999

Year period	Victoria	South Australia	Tasmania	Northern Territory	Total
1970–1974	7/353,503 (2.0)	3/107,557 (2.8)	0/39,197 (0)	1/14,157 (7.1)	11/514,414 (2.2)
1975–1979	20/297,750 (6.7)	4/94,855 (4.2)	0/34,135 (0)	2/13,112 (15.2)	26/439,852 (5.9)
1980–1984	31/296,595 (10.5)	8/96,782 (8.3)	3/35,239 (8.5)	0/14,943 (0)	42/443,559 (9.5)
1985–1989	44/309,360 (14.2)	17/97,531 (17.4)	2/34,581 (5.8)	1/16,977 (5.9)	64/458,449 (14.0)
1990–1994	30/326,197 (9.2)	23/98,301 (23.4)	5/34,579 (14.5)	1/18,127 (5.5)	59/477,204 (12.4)
1995–1999	34/303,822 (11.2)	8/92,815 (8.6)	2/31,033 (6.4)	1/20,559 (4.9)	45/448,229 (10.0)
Total	166/1,887,227 (8.8)	63/587,841 (10.7)	12/208,764 (5.7)	6/97,875 (6.1)	247/2,781,707 (8.9)

Several centers have reported that mortality after the Fontan procedure is now very low.[3,4] Children having the Fontan procedure in the 1970s had a very poor long-term outcome: half of them died or required transplantation in the 25 years after the Fontan procedure.[5] After the introduction of improved techniques, patients now have a better chance of symptom-free survival for more than 20 years. Knowing the incidence of Fontan surgery in the population may help to estimate the future size of the Fontan population entering adulthood. For example, it is likely that, with 250,000 births a year, Australia will have 25 new patients undergoing a Fontan procedure each year. These patients are likely to need intensive medical support, and perhaps heart transplantation, in the decades to come.

Longer survival, the growing spectrum of conditions necessitating a Fontan circulation, the inevitable late morbidity and mortality, and the requirement for specialist intervention when failure occurs all point to a need to plan for the care of a growing number of adults who have had Fontan surgery.

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